JACC Vol. 7, No. 5
May 1986:1104–6

Evaluation of Pulmonary Vascular Resistance in the Presence of a Large Pulmonary Arteriovenous Malformation

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A 1 day old infant presented with severe cyanosis and congestive heart failure. Cardiac catheterization confirmed the clinical suspicion of a pulmonary arteriovenous malformation of the right lower lobe. Catheter occlusion of the right lower pulmonary artery allowed evaluation of the integrity of the remaining pulmonary vascular bed as well as improvement of the patient's hemodynamic condition pending more definitive therapy.

(J Am Coll Cardiol 1986;7:1104–6)

Pulmonary arteriovenous malformations have been recognized as a rare cause of cyanosis and heart failure in the newborn infant (1–3). When the malformation is single, surgical therapy is indicated and is generally successful (2). However, diagnosis of a large malformation that steals blood from most of the lung, resulting in both cyanosis and heart failure on the first day of life, fails to establish the integrity of the rest of the pulmonary vascular system. Such a situation has been associated with marked hypoplasia of the remaining pulmonary arteries (4). Measurement of pulmonary artery pressure and pulmonary vascular resistance in the presence of a large, low resistance pulmonary arteriovenous malformation may severely underestimate the true pulmonary vascular resistance in the remaining, potentially normal lung. We describe our assessment of such a newborn and the usefulness of catheter occlusion of the arteriovenous malformation in assessing these variables and improving the infant's clinical status before definitive therapy.

Case Report

A 1 day old male infant was admitted to our intensive care nursery with cyanosis and heart failure. He appeared active and alert but had marked central cyanosis. The heart rate was 150 beats/min, respiratory rate 80/min and blood pressure 70/40 mm Hg. The liver was palpated 5 cm below the right costal margin and the precordium was mildly hyperdynamic with the apical beat shifted 2 cm lateral to the midclavicular line in the sixth intercostal space. There were no murmurs. On chest X-ray film the cardiothoracic ratio was 0.64, and the lung fields were clear except for an opacity in the right lower lobe (Fig. 1). An arterial blood gas determination (FIO_2 = 1.0) showed a pH of 7.4, partial pressure of oxygen (Po_2) of 30 mm Hg, partial pressure of carbon dioxide of 36 mm Hg and bicarbonate of 22 mmol/liter. An echocardiogram demonstrated a structurally normal heart with a patent foramen ovale. The patient's oxygenation did not improve with ventilation.

The diagnosis of pulmonary arteriovenous malformation was considered and confirmed at cardiac catheterization. The hemodynamic data (Table 1) demonstrated a pulmonary arteriovenous malformation isolated to the right lower lobe of the lung. The pulmonary to systemic systolic artery pressure ratio was 0.44. Right ventricular and right pulmonary artery angiography demonstrated the large arteriovenous malformation of the right lower pulmonary lobe (Fig. 2). Because of the massive flow of blood through this area, the remaining pulmonary vascular bed was poorly perfused and inadequately visualized, suggesting functional or structural hypoplasia, or both. A 5F Berman angiographic catheter was then passed into the right lower pulmonary artery, and with balloon occlusion of this vessel the systemic arterial Po_2 rose to 210 mm Hg while the pulmonary artery pressure remained low. A hand injection of contrast material (Fig. 3) demonstrated virtually complete occlusion of the right lower pulmonary artery and showed adequate caliber of the pulmonary vessels supplying the remaining lung. The balloon was filled with radiopaque solution and left inflated. A chest X-ray film 1 hour later demonstrated a marked

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Manuscript received November 12, 1985; accepted December 20, 1985.

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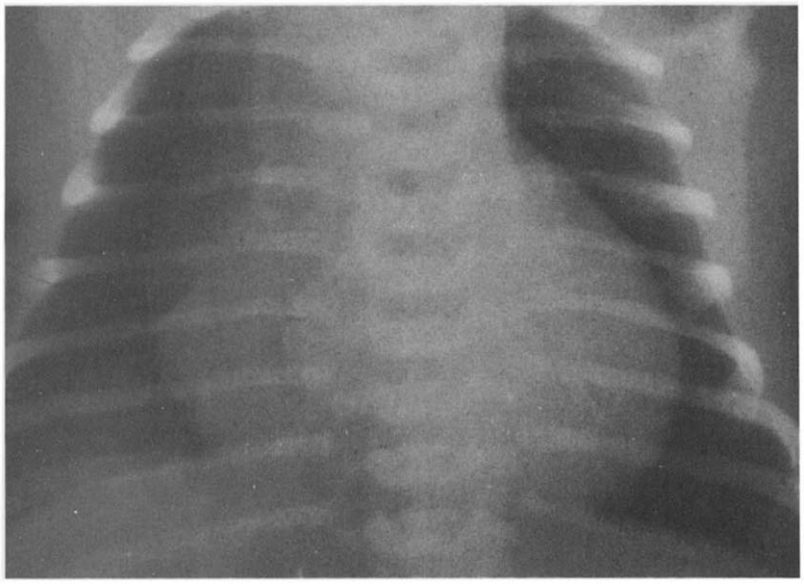

Figure 1. Chest X-ray film on hospital admission. The infant is slightly rotated. The cardiac silhouette is enlarged with a cardiothoracic ratio of 0.64. An opacity is evident in the right lower lobe of the lung.

decrease in the heart size with a cardiothoracic ratio of 0.51 (Fig. 4).

At operation, lobectomy of a virtually bloodless right lower lobe was carried out without complication. In the first 36 hours postoperatively the pulmonary to systemic systolic arterial pressure ratio remained less than 0.5. At discharge at 6 days of age the infant was fully saturated while breathing room air.

Discussion

Catheter occlusion of the right lower pulmonary artery was useful in evaluating pulmonary artery pressure and pulmonary vascular resistance once flow through the arteriovenous malformation was effectively removed so as to permit preoperative evaluation of the integrity of the uninvolved pulmonary vascular bed. Also, angiographic demonstration

Table 1. Hemodynamic Data and Pulmonary Artery and Aortic Pressures

Hemodynamic Data*			
	Pressure (mm Hg)	Oxygen Saturation (%)	P_{O_2} (mm Hg)
Superior vena cava		55	
Inferior vena cava		68	
Right atrium	*a* 8; *v* 11, m 6	71	
Right ventricle	40/6		
Main pulmonary artery	36/10; m 24	67	
Left lower pulmonary vein		95	484
Right lower pulmonary vein		61	28
Right upper pulmonary vein		95	500
Left atrium	*a* 6; *v* 10, m 6		
Left ventricle	90/8	81	
Aorta	88/68; m 76	81	35

Simultaneous Pulmonary Artery and Aortic Pressures†			
	Main Pulmonary Artery (mm Hg)	Aorta (mm Hg)	Systolic Pressure Ratio
Before balloon occlusion	36/10	82/64	0.44
Balloon occlusion (10 min)	52/20	110/74	0.47
Balloon occlusion (20 min)	38/16	92/68	0.41

*Hemodynamic data obtained at the time of cardiac catheterization; concentration of inspired oxygen in air (FIO_2) = 1.0. †The simultaneous pulmonary artery and aortic pressures and the systolic pressure ratio before and at 10 and 20 minutes after balloon occlusion of the right lower pulmonary artery. *a* = *a* wave; m = mean; *v* = *v* wave.

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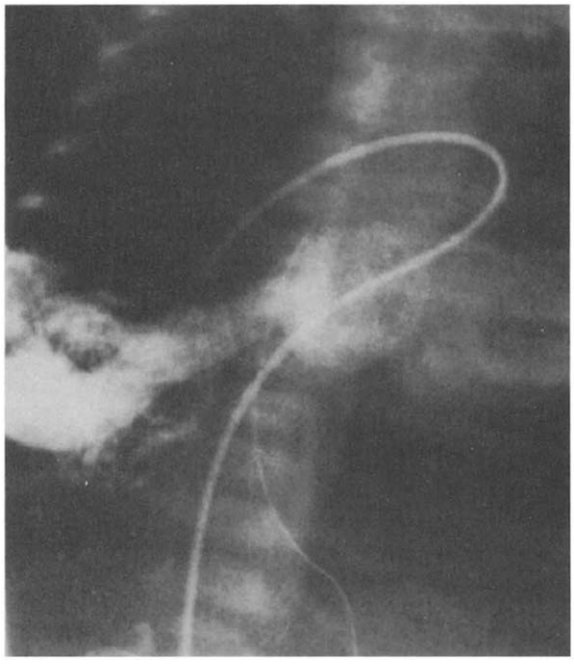
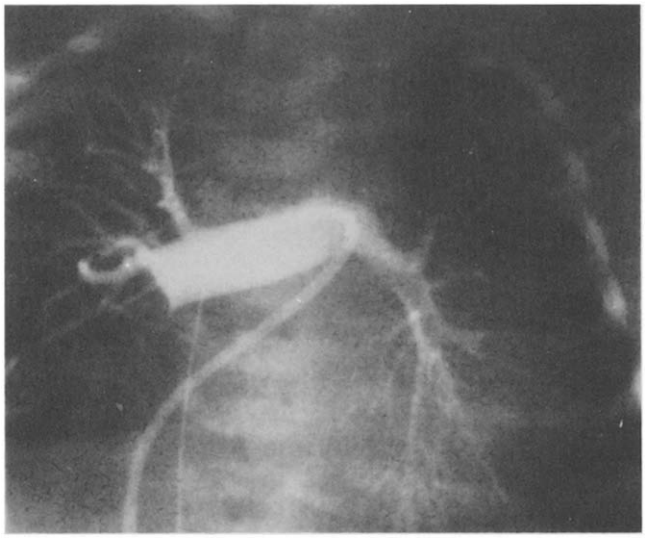


Figure 2. Venous phase of a right pulmonary artery angiogram demonstrating the large arteriovenous malformation of the right lower lobe. Neither a right ventricular angiogram nor the arterial phase of this angiogram demonstrated adequate pulmonary arteries to the right upper lobe or left lung.

Figure 3. Right pulmonary artery arteriogram after balloon occlusion of the right lower pulmonary artery. Normal arterial vessels are seen in the right upper and middle lobes as well as a normal left pulmonary artery with filling of the left lower lobe. The left upper lobe was not visualized.



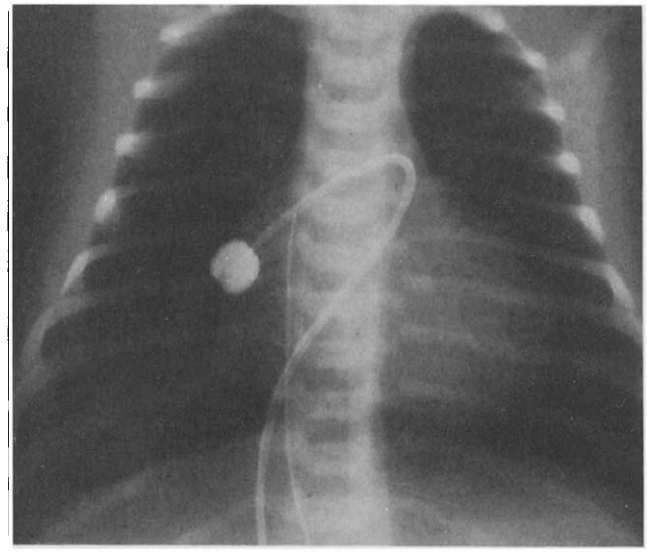


Figure 4. Chest X-ray film taken 1 hour after balloon occlusion of the right lower pulmonary artery. The balloon was inflated with a solution containing contrast material. The cardiac silhouette has markedly decreased in size from the previous examination and the cardiothoracic ratio is 0.51.

of the pulmonary vessel morphology, which could not be visualized adequately while the malformation was open and stealing blood from the rest of the lung, was obtained. Finally, occlusion of the arteriovenous malformation before surgery resulted in improved oxygenation and diminished right to left shunt and volume overload on the heart. Therefore, whether surgery or embolization is the planned treatment for this condition, this technique can be used to assess the anticipated functional status after removal of the malformation.

We thank Mary Hornby for assistance in the preparation of this manuscript.

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